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Natural history of non-surgical complete atrioventricular block in children and predictors of pacemaker implantation

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Objectives:
Data on natural history of non-surgical complete atrioventricular block (CAVB) in children are scarce and criteria for pacemaker implantation (PM) are based on low level of evidence. We aimed to evaluate natural course and predictors of PM in a nation-wide cohort of paediatric patients with CAVB.

Methods:
All paediatric patients presenting between 1977 and 2016 with CAVB in absence of any but trivial structural heart disease, were retrospectively identified from the institutional database yielding 95 subjects (female 54, male 41) aged median 4.06 (IQR 0.10-10.34) years at first presentation. Patients were followed-up for median 0.80 (IQR 0.02-7.07) years providing a total of 347 patient-years available for analysis until PM or end of follow-up. Serial 24-hour Holter recordings and echocardiograms were reviewed to asses heart rate profiles, left ventricular (LV) size and shortening fraction. Absolute values and z-scores of healthy population were used for analysis. Predictors of PM performed >1 month after first presentation were evaluated using Cox PH model.

Results:
Absolute minimum and mean 24-hour heart rates and maximum RR intervals had a non-linear correlation with age ($p<0.0001$ for all) with maximum progression of bradycardia during the first 2 years of life. Both LV end-diastolic diameter (median 1.49, IQR 0.27-2.67 z) and shortening fraction (median 36, IQR 31-43 %) at presentation were stable throughout follow-up. PM was performed in 64 patients (67.4 %) reaching published guidelines criteria. Probability of freedom from PM was 56.9/45.3/42.4/29.7 % at 1/3/5/10 years. Mean heart rate at presentation was the strongest predictor of PM (HR=0.939, CI 0.894-0.986, $p=0.011$ per unit increase) regardless of presentation age. Patients presenting with a mean heart rate >58 BPM (>75 centile of the group) had high probability of freedom from PM within the subsequent 5 years (91.7 %) as opposed to rest of the group (41.2 %), $p=0.012$. Echocardiographic parameters did not predict PM.

Conclusions:
Paediatric patients with CAVB show an age-dependent decrease in heart rates, stable degree of LV remodelling and preserved LV function. Need for PM can be predicted by the heart rate profile at presentation defining a low risk group and allowing for stratified follow-up.